ACTA SCIENTIFIC CANCER BIOLOGY

Volume 3 Issue 11 November 2019

Mini Review

Non-Langerhans Cell Histiocytoses (Non-LCH)

Behzad Saberi*

Medical Research, Esfahan, Iran

***Corresponding Author:** Behzad Saberi, Medical Research, Esfahan, Iran.

Received: October 14, 2019; **Published:** October 23, 2019

Abstract

Non-Langerhans Cell Histiocytoses or Non-LCH is a group of different pathologies with specific clinical manifestations. This study is a brief review on some of those pathologies in the Non-LCH category, which may show some involvements of the CNS in their presentation.

Keywords: Non-Langerhans Cell Histiocytoses; CNS Involvement.

Introduction

Non-Langerhans Cell Histiocytoses or Non-LCH is a group of lesions which show no langerhans cell differentiation but macrophage. Familial hemophagocytic lymphohistiocytosis, Erdheim-Chester disease, Xanthoma disseminatum, Jevenile Xanthogranuloma, Choroid plexus xanthogranuloma and Rosai-Dorfmann disease are the pathologies in the Non-LCH category, which may show the involvement of the CNS.

Familial hemophagocytic lymphohistiocytosis as a rare autosomal recessive disorder, presents with high immune activation. Macrophages and T-lymphocytes affect various organs including CNS and cause multisystem disorder of infancy with a rapid and progressive manner. If bone marrow transplantation would not be done for treatment, the average survival would be about two months. Germ line mutations in some genes like MUNC13-4 on 17q25 and PRF1 on 10q22 would cause the disease.

Xanthoma disseminatum is associated with generalized hyperlipidemia. It is related to the lesions composed of lipidized histiocytes. Dura mater or pituitary and hypothalamic regions would be the locations of CNS involvement in this pathology.

Erdheim-Chester disease can involve multiple organs specifically in adults. Meninges, cerebellum and pituitary region are the most common locations of the intracranial lesions in this disease.

Juvenile xanthogranuloma presents itself as skin nodule formation in children group. Visceral involvement of the meninges and the brain can also be seen.

Choroid plexus xanthogranuloma as lesions which are located in the brain ventricles specifically in the lateral ventricles, in some rare cases may cause obstruction in the CSF flow. Foreign-body giant cells, foaming macrophages, cholesterol clefts and chronic inflammatory infiltrates compose the granulomatous lesion in the choroid plexus xanthogranuloma.

In choroid plexus xanthoma, only foamy macrophages can be seen.

Rosai-Dorfmann disease would be presenting as an intracranial and dural-based mass which can be similar to a meningioma. Resection of the mass makes Rosai-Dorfmann disease prognosis good. Only in some of the patients with intracranial lesions the clinical features of the disease like weight loss, fever and cervical lymphadenopathy can be seen [1-9].

Conclusion

Having knowledge about the Non-Langerhans Cell Histiocytoses or Non-LCH pathologies specifically ones with CNS involvement, is of great importance to diagnose the relevant patients with such pathologies and make proper decisions to treat such patients effectively during clinical practice.

Bibliography

1. Weidauer Stefan., *et al*. "Cerebral Erdheim-Chester disease: Case report and review of the literature". *Neuroradiology* 45 (2003): 241-245.

2. Haroche J., *et al*. "Erdheim-Chester disease". *Current Rheumatology Reports* 16 (2014): 412.

3. Emile JF., *et al*. "Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages". *Blood* 127 (2016): 2672-2681.

4. Cypel., *et al*. "Juvenile Xanthogranuloma: Case Report and Review of the Literature". *The Canadian Journal of Plastic Surgery* 16.3 (2008): 175-177.

5. Alexander AS., *et al*. "Xanthoma disseminatum: a case report and literature review". *British Journal of Radiology* 78 (2005): 153-157.

6. Cai Y., *et al*. "Review of Rosai-Dorfman disease: new insights into the pathogenesis of this rare disorder". *Acta Haematology* 138 (2017): 14-23.

7. Usmani G Naheed., *et al*. "Advances in understanding the pathogenesis of HLH". *British Journal of Haematology* 161 (2013): 609-622.

8. Benson LA., *et al*. "Pediatric CNS-isolated hemophagocytic lymphohistiocytosis". *Neurology Neuroimmunology and Neuroinflammation* 6 (2019): e560.

9. Weitzman S and Jaffe R. "Uncommon histiocytic disorders: the non-Langerhans cell histiocytoses". *Pediatric Blood Cancer* 45 (2005): 256-264.

Volume 3 Issue 11 November 2019